CLINICAL CASE
doi: 10.35366/117372

Cirujano General
January-March 2024
Vol. 46, no. 1 / p. 73-80

Type IVA common bile duct cyst based on Todani classification

Quiste de colédoco tipo IVA basado en la clasificación de Todani

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Keywords: common bile duct cyst, Todani classification, cholangioresonance, Roux-en-Y biliary reconstruction.

Palabras clave: *quiste de colédoco, clasificación de Todani, colangiorresonancia, reconstrucción biliar en Y de Roux.*

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Received: 08/03/2023
Accepted: 07/22/2024

ABSTRACT

Cystic dilatations of the common bile duct are a rare congenital anomaly that allows bile reflux, causing dilatation of the bile ducts in various forms. In the last five years (2018-2022), the general surgery service of the Civil Hospital of Guadalajara "Fray Antonio Alcalde" attended a total of 5,886 patients, among which only two cases of choledochal cyst (premalignant lesions) were found, with an incidence rate of 0.03%, presenting a low frequency of this pathology in our region compared to western nations and an even lower incidence concerning the Asian population. We present the case of a 36-year-old female patient from the state of Hidalgo with a biliary cystic lesion staged as IVA, according to the modified Todani classification, with a magnetic cholangioresonance study. After a complete preoperative evaluation, the cystic lesion was resected, and the biliary tract was reconstructed with jejunal and Roux loops; she evolved satisfactorily and was discharged on the eighth postoperative day. She was seen in the outpatient clinic with hepatic functional tests and a new magnetic cholangioresonance reported as normal and with good biliary drainage.

RESUMEN

Las dilataciones quísticas del colédoco son una anomalía congénita infrecuente que permite el reflujo de bilis, ocasionando la dilatación de las vías biliares en diversas formas. En los últimos cinco años (2018-2022) el servicio de cirugía general del Hospital Civil de Guadalajara "Fray Antonio Alcalde" atendió un total de 5,886 pacientes entre los cuales se encontraron sólo dos casos de quiste de colédoco (lesiones premalignas), con una tasa de incidencia de 0.03%, presentando una baja frecuencia de esta patología en nuestra región en comparación con naciones occidentales y una incidencia aún más baja en relación a la población asiática. Se presenta el caso de una femenina de 36 años derivada del estado de hidalgo con una lesión quística biliar, la cual se estadificó como IVA, de acuerdo con la clasificación modificada de Todani, con estudio de colangiorresonancia magnética. Previa valoración completa preoperatoria se resecó la lesión quística y se reconstruyó la vía biliar con asas de yeyuno en Y de Roux; evolucionando de forma satisfactoria y siendo dada de alta al octavo día postoperatorio, vista en consulta externa con pruebas funcionales hepáticas y nueva colangiorresonancia magnética cuyo reporte resultó completamente normal y con buen drenaje biliar.

INTRODUCTION

The first description of cystic dilatation of the biliary tree was attributed to Vater in 1720. Subsequently, several reports have referred to cases of patients with the triad of jaundice, pain, and abdominal mass suggestive of cystic lesions of the biliary tract.[1]

Choledochal cysts have an incidence in Western countries of 1:13,500 and in Asian populations of 1:1,000, with type IV being the most frequent in adults.[2] In addition, it is

How to cite: Ocampo-González S, Martínez-Partida JI, Rodrigo-Pozo S, Ocampo-Barro SA, Rea-Vázquez IA, Cuevas-Sánchez MT. Type IVA common bile duct cyst based on Todani classification. Cir Gen. 2024; 46 (1): 73-80. https://dx.doi.org/10.35366/117372

suggested that the prevalence in both Western and Eastern women is 4:1.[1]

According to the literature, the most accepted theory regarding etiology is the presence of an anomalous pancreaticobiliary junction that allows reflux and a consequent increase in pressure that causes dilatation, inflammation due to activation of pancreatic enzymes, biliary cholestasis, and epithelial damage.[3]

Clinical manifestations vary between adults and infants, and although most cases of common bile duct cysts are diagnosed during childhood, 25% are discovered in adulthood. Abdominal pain, palpable abdominal mass, and jaundice, also known as the classic triad of common bile duct cysts, are found in only 20% of cases. Adults often present with nonspecific symptomatology, such as right upper quadrant abdominal pain, jaundice, nausea, vomiting, and fever.[4]

Diagnosis is based on studies, including blood cytology, electrolytes, liver function tests, International Normalized Ratio (INR), and tumor markers. In addition, abdominal ultrasound (USG) allows visualization of hepatic and pancreatic structures. However, endoscopic retrograde cholangiopancreatography (ERCP) and magnetic cholangioresonance (M-CR) have become the gold standard for proper diagnosis and classification.[5,6]

In 1959, Alonso-LEJ and collaborators attributed the term common bile duct cyst. They classified them into three types, among which the following stand out: congenital cystic dilatations, congenital diverticulum of the common hepatic (CH) duct, and congenital choledochococele.[5]

Later, in 1977, Todani and associates added two more types and several subtypes to the classification, being updated by the same authors in 1997 and 2003.[7,8] The most recent Todani classification subdivides type I being the most frequent (> 90%); variation IA involves dilatation of the entire extrahepatic bile duct, unlike type IB which includes dilatation of a segment of the common bile duct below the cystic implantation and type IC comprises a fusiform or cylindrical dilatation of the entire extrahepatic common bile duct.[7,8] Finally, subtype ID was proposed and added to the Todani classification modified in 2008 by Calvo-Ponce and colleagues, which is a segmental dilatation above the cystic and bifurcation of the hepatic ducts *(Figure 1)*.[9]

In type II, there are diverticula throughout the extrahepatic duct. Type III is a choledochocele, and type IVA has the presence of multiple cysts in the intrahepatic and extrahepatic bile duct. In type IVB, multiple cystic dilatations of the extrahepatic bile duct are present. Type V (Caroli's disease) involves dilatation of one

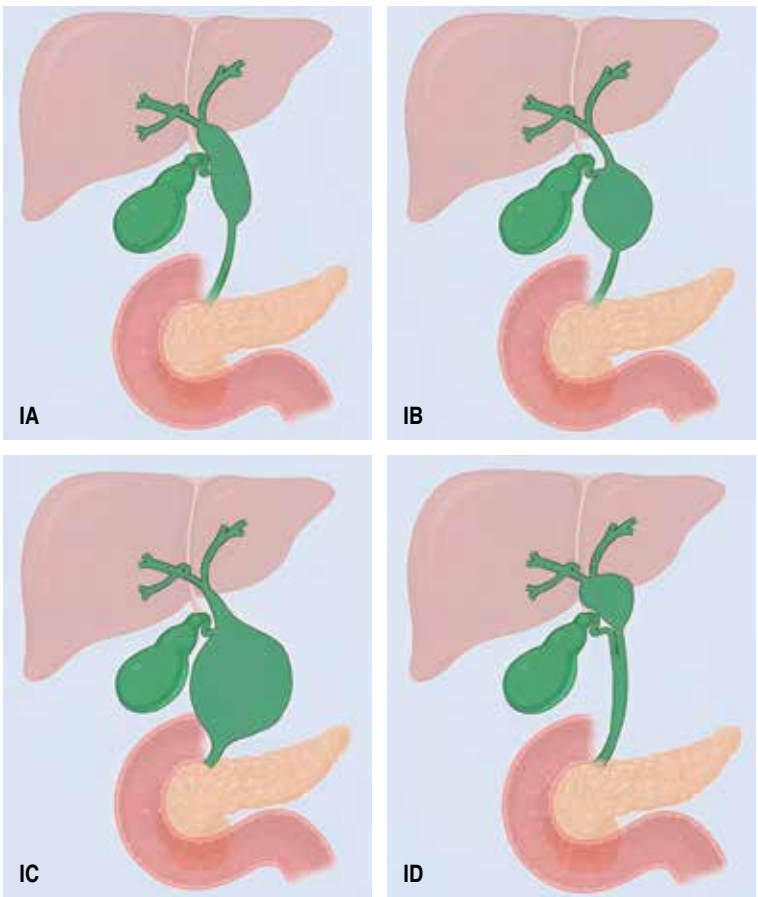

Figure 1: *Subclassifications of modified Todani type I common bile duct cyst. Subclassifications of type I choledochal cyst:* ***(IA)*** *cystic dilatation involves the entire extrahepatic biliary system,* ***(IB)*** *involves only a portion of the extrahepatic bile duct,* ***(IC)*** *spindle-shaped expansion of the whole of extrahepatic bile ducts, along with widening of the intrahepatic ducts,* ***(ID)*** *extrahepatic fusiform dilatation at the biliary confluence, with a non-dilated intrahepatic biliary tree and no pancreatobiliary malunion.*[2]

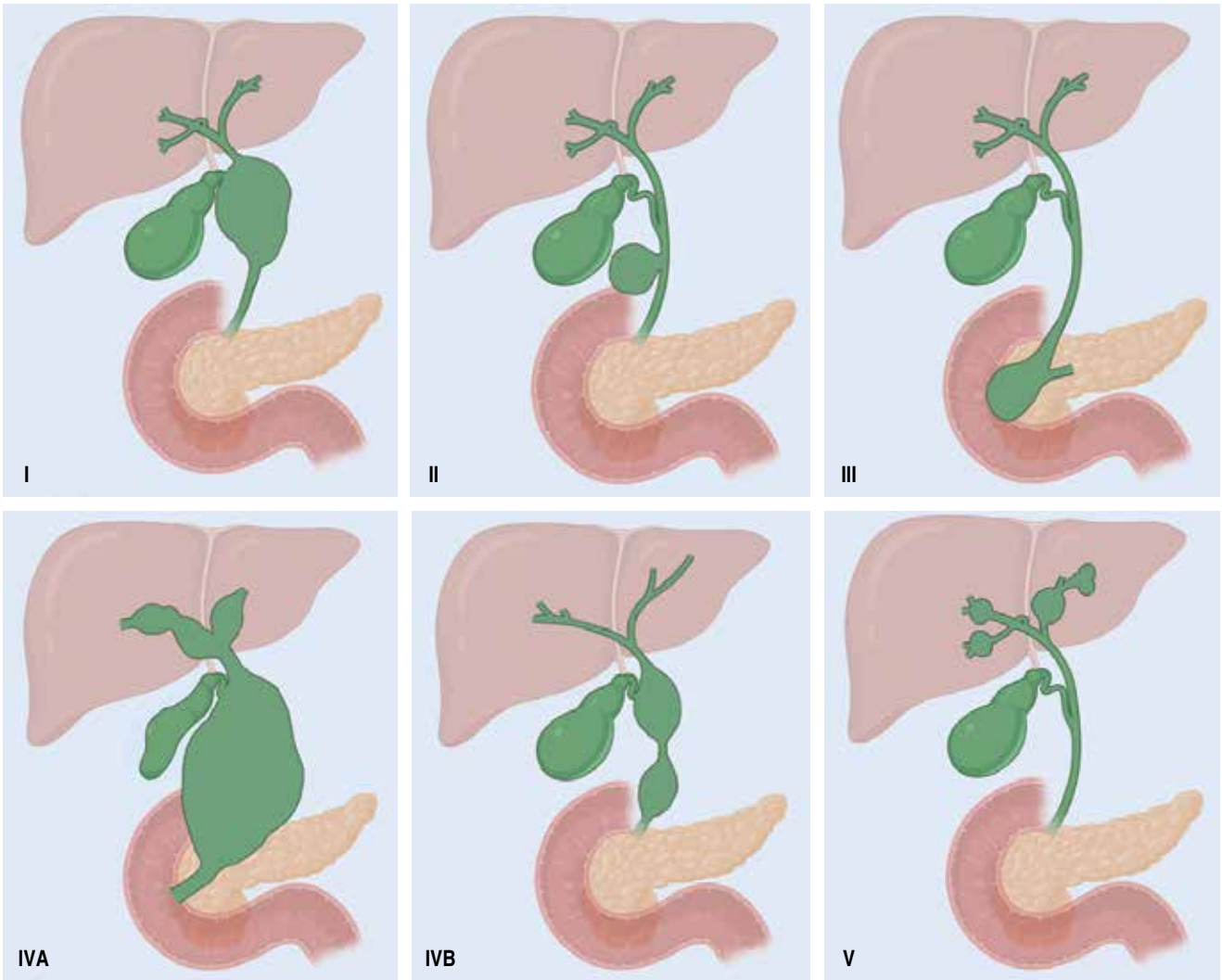

Figure 2: *Modified Todani classification:* ***(I)*** *subclassified in four ways mentioned in Figure 1,* ***(II)*** *corresponds to a diverticular sac in the common bile duct,* ***(III)*** *also called choledochococele, consisting of an intramural dilatation of the distal segment of the common bile duct,* ***(IVA)*** *consists of dilatation of the bile ducts both inside and outside the liver,* ***(IVB)*** *exclusively extrahepatic bile ducts show expansion at different points,* ***(V)*** *existence of single or multiple dilatations that are limited to the bile ducts within the liver, also called Caroli's disease.*[2]

or several segments of the intrahepatic ducts *(Figure 2)*.[5,10]

In 2004, Visser and associates proposed a modification and distinguished the following designations: common bile duct cyst, diverticulum, and Caroli's disease.

In 2011, Michaelides and collaborators reported and proposed a new variant called type ID; in this new variant, in addition to the dilatation of the HC, there is a dilatation of the central portion of the cystic duct, giving a bicornuate configuration to the cyst.[2]

The treatment of choice is surgical resection, avoiding drainage of the cyst or incomplete resection, especially of the mucosa of the affected segment of the bile duct to prevent dysplasia and postoperative malignant degeneration,[11] since common bile duct cysts are precancerous lesions with a malignancy rate that ranges from 2.5 to 28%, increasing with age.[2]

PRESENTATION OF THE CASE

A 36-year-old woman, originally from the state of Hidalgo, Mexico, underwent surgery at the general surgery service of the Hospital Civil de Guadalajara "Fray Antonio Alcalde" (HCGFAA) in Jalisco, Mexico, on November 28, 2022, due to the presence of a common bile duct cyst type IVA according to the modified Todani classification (QC-IVA-Todani-modified). She had an evolution of two months characterized by epigastric abdominal pain, early satiety, and anorexia without nausea, vomiting, fever, or jaundice.

In her home, a hepatic and biliary tract ultrasound (HBVUS) was performed to interpret the presence of a hepatic cyst. Later, an M-RC was performed in Pachuca, Hidalgo, which confirmed the general surgeon's diagnosis of a common bile duct cyst. Due to a lack of resources, the surgeon referred her to the HCGFAA. Laboratory tests were performed *(Table 1)*, and a new CR-M was performed *(Figures 3 and 4)*, which showed a modified QC-IVA-Todani cyst *(Figure 2)*.

Findings: an atrophic gallbladder without the presence of gallstones, a 7.6 mm cyst in the cystic duct, saccular dilatation of the intrahepatic and extrahepatic bile duct with a diameter of 18 mm in the right hepatic, 15 mm in the left hepatic duct and a confluence

Table 1: Pre- and post-surgical laboratory tests and six-month follow-up.

Laboratory tests	Pre-surgical	Post-surgical	Six-month follow-up
Blood cytology	Hemoglobin: 14.3 g/dl Platelets: 185,000 mm^3 Leukocytes: 5.3 Neutrophils: 2.7	Hemoglobin: 16.6 g/dl Platelets: 174,000 mm^3 Leukocytes: 20.5 Neutrophils: 18.5	Hemoglobin: 13.6 g/dl Platelets: 130,000 mm^3 Leukocytes: 7.0 Neutrophils: 62% Htc: 41.8% MCV: 91 FI MCHC: 31 pg Lymphocytes: 32% Monocytes: 3% Basophils: 0% Eo: 3% Segmented: 62% Bands: 0%
Blood chemistry	Glucose: 95 mg/dl Urea: 13 mg/dl Creatinine: 0.58 mg/dl	Glucose: 75 mg/dl Urea: 23 mg/dl Creatinine: 0.51 mg/dl	Glucose: 102 mg/dl Urea: 29 mg/dl Creatinine: 0.78 mg/dl
Liver function tests	GGT: 23 U/l AST: 20 U/l ALT: 16 U/l TB: 0.28 mg/dl DB: 0.04 mg/dl IB: 0.24 mg/dl	GGT: 194 U/l AST: 19 U/l ALT: 29 U/l TB: 0.54 mg/dl DB: 0.17 mg/dl IB: 0.37 mg/dl	GGT: 24 U/l AST: 30 U/l ALT: 25 U/l TB: 0.50 mg/dl DB: 0.20 mg/dl IB: 0.30 mg/dl

ALT = alanine aminotransferase. AST = aspartate aminotransferase. DB = direct bilirubin. IB = indirect bilirubin. TB = total bilirubin. MCHC = mean corpuscular hemoglobin concentration. Eo = eosinophils. GGT = gamma-glutamyl transpeptidase. Hct = hematocrit. MCV = mean corpuscular volume.

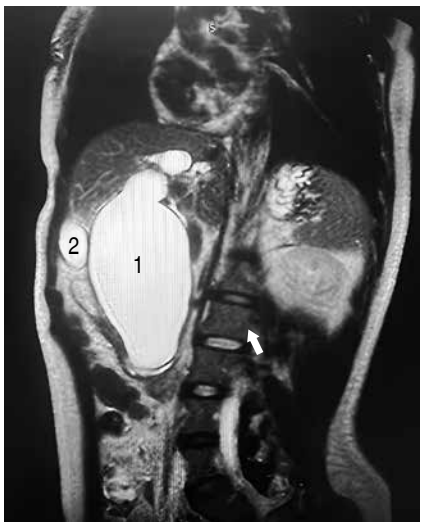

Figure 3: *Magnetic resonance cholangiography. Coronal section of magnetic resonance cholangiography; (1) common bile duct cyst (70 mm) with interspersed areas of stenosis and added intrahepatic dilatation that classifies it as type IVA. (2) Normal-sized gallbladder with a thin wall.*

zone of 33 mm (carina) was seen. A maximum dilatation of the common bile duct of 70 mm, distal common bile duct of 23 mm, and the duct of Wirsung with normal dimensions were observed.

Elective resection was scheduled laparoscopically *(Figure 5)* after completing a thorough preoperative protocol. Intraoperative findings evidenced a QC-IVA-Todani-modified, with an atrophic gallbladder. Dissection of the cyst capsule was initiated by attempting to separate it from the vascular structures (portal vein and hepatic artery). It was decided to open the cyst to facilitate its dissection. However, due to its strong adhesion and vascularization, with scarce but continuous bleeding, it was converted to open surgery through a Kocher-type incision.

Dissection of the cyst was continued by separating it from the portal vein without damaging it. A Bakes dilator was introduced through the cyst opening to identify and canalize the dilated right and left hepatic ducts. Then, the removal of the common bile duct cyst was performed from the bifurcation of both intrahepatic ducts to the duodenal border, where a continuous suture with polydioxanone was performed, including the last centimeter of the common bile duct, thus performing a complete removal of the cyst, including the gallbladder. Finally, a Roux-en-Y biliodigestive bypass was performed, with a jejunal loop of 50 cm from the Treitz angle and a biliary drainage loop of 50 cm, using a 60 mm linear stapler in the hepatic-jejunal anastomosis.

The jejunum-jejunal anastomosis was performed manually with a continuous suture of Vicryl® 4-0, leaving an anastomotic diameter of 30 mm. Hemostasis was verified; a gauze count was performed, and a closed drain of 19 Fr was installed. A bleeding of 800 cc was recorded, so a globular package was transfused during the trans-operative period. The patient was given antibiotic therapy with ceftriaxone 1 g every/12 hours and metronidazole 500 every 8 hours. On the following postoperative day, she started on a liquid diet and ambulation, and on the third postoperative day, she was given a mixed diet.

The cyst was sent to pathology evaluation with the following findings: a cyst wall

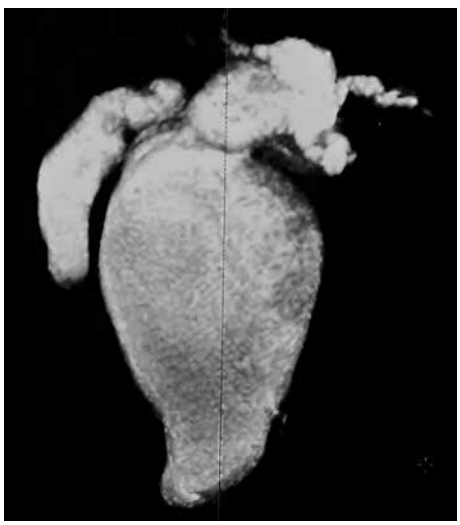

Figure 4: *Magnetic resonance cholangiography. 3D reconstruction of magnetic resonance cholangiography: widening of the common bile duct up to 70 mm, dilatation of the right hepatic duct with 18 mm and 15 mm of the left hepatic duct, and 30 mm in the confluence zone.*

composed of dense fibroconnective tissue lined by simple columnar epithelium accompanied by acute and chronic inflammatory infiltrate. A lamina propria with vasodilatation and acute and chronic infiltrate extending to the cyst wall was seen. Focal papillary hyperplasia of the mucosa, areas of focal ulceration, and a secondary lymphoid follicle formation *(Figure 6)* were shown.

The patient had a good clinical evolution and was discharged on postoperative day 8 with no evidence of early surgical complications. Laboratory studies continue to be performed.

There were six-month follow-up appointments with laboratory tests, including blood cytology, blood chemistry, and liver profile, which found the patient clinically stable and physical examination without jaundice, cardiopulmonary problems, and abdomen with good surgical healing process. The patient will continue to be followed up with a CT scan with intravenous water-soluble contrast and determination of tumor markers: carcinoembryonic antigen (CEA) and CA 19-9 at 12 months post-surgery.

DISCUSSION

In the last five years, the general surgery service of the HCGFAA attended 5,886 patients, of which only two cases of common bile duct cysts were found in adults. The incidence of this pathology in HCGFAA was 0.03%, low compared to Western countries and even lower concerning the Asian population. Choledochal cysts are more commonly found in Eastern countries, with a prevalence of 1:1,000 persons, with type IV being the most frequent in adults.[2] The sex ratio is reported to be more frequent in women in both Eastern and Western countries, 4:1.[1]

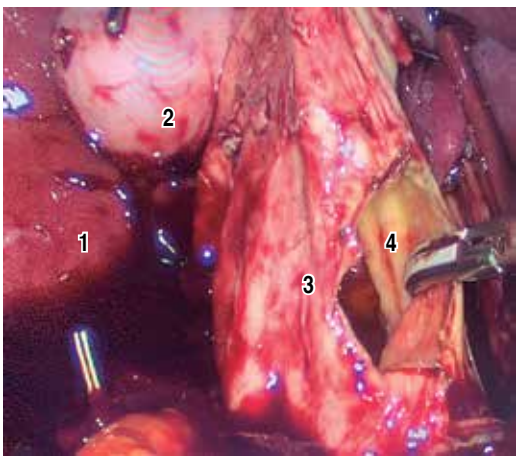

Figure 5: *Laparoscopic view. Opening of the common bile duct cyst at its anterior wall; (1) right hepatic lobe; (2) gallbladder; (3) anterior serosa wall of the cyst; (4) posterior mucosa wall of the cyst.*

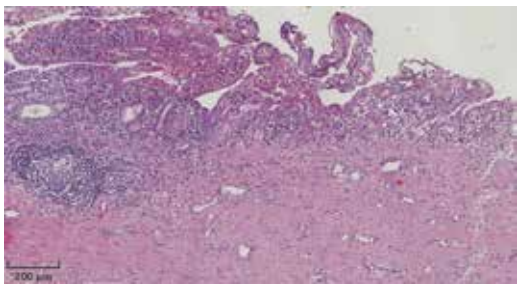

Figure 6: *Laminoma. Focal papillary hyperplasia of the mucosa, with chronic and acute inflammatory infiltrate, areas of focal ulceration. Formation of a secondary lymphoid follicle.*

Due to the inherent pathophysiology of common bile duct cysts, the scientific literature mentions a malignancy rate that varies between 2.5 and 28%, which increases with age.

The variety of common bile duct cysts leads us to divide their management according to their classification; surgical resection for type I and IV cysts is the therapeutic option of choice, either by laparoscopic, robotic, or open surgery. The use of the laparoscopic and robotic routes entails a considerably lower risk of both immediate and late complications, according to Miron et al.[2] Among the most common surgical complications are bleeding and dehiscence of the anastomosis, which could result in the appearance of leaks leading to local or generalized sepsis of the abdominal cavity. This situation requires immediate treatment according to the current protocols for managing the dehiscence in biliodigestive anastomoses. For type II cysts, diverticulectomy plus primary closure of the common bile duct can be chosen. If necessary,

management can be limited to transduodenal excision for type III cysts by endoscopic management with respective sphincterotomy and sphincteroplasty.

However, due to the firm adhesion of the posterior wall of the cyst to the vascular structures, mainly to the portal vein, laparoscopic resection was not possible, and it became an open surgery through a Kocher-type incision. In this way, its total extirpation was achieved, leaving a remnant of the duct in the intrapancreatic portion of about 5 mm. The literature indicates that this remnant can be the cause of dysplasia.[11,12] However, we do not fully agree with this statement because the physiopathology of the cyst is modified when most of the cystic lesion is removed, thus eliminating the increase of pressure in the biliary tract.

Zheng and colleagues debated whether VATS cysts in adults require additional hepatic resection because of concomitant intrahepatic dilatation; in this case, hepatic resection is unfeasible since the reported dilatations involve the initial portion of both hepatic ducts. Therefore, resection should be limited to specific segments that do not increase the morbimortality of the patient due to extensive hepatic resection. This situation is replicated in pediatric patients since they have a much milder inflammatory evolution and less malignant transformation than adults, less lithiasis formation, and a lower risk of cholangiocarcinoma in general.[10]

In the present case, the patient was followed up for 10 months. Her evolution will continue to be monitored by CT scan with water-soluble intravenous contrast and determination of tumor markers: carcinoembryonic antigen (CEA) and CA 19-9 every six months until five years postoperatively, to ensure the non-recurrence or malignant transformation of the unresected cystic remnant.

CONCLUSIONS

Giant common bile duct cyst, a rare condition worldwide, may present with nonspecific symptoms, leading to its diagnosis going unnoticed, highlighting the importance of resorting to imaging techniques such as ERCP or M-CR for timely evaluation and staging. In addition, long-term postoperative monitoring is required to prevent complications, particularly malignant transformation. The latter can affect the bile ducts, gallbladder, and even the pancreas in advanced stages, decades after the initial intervention.

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